# What's New in Cerebral Palsy

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*• Among new researches bearing on cerebral palsy are the growth of brain cells in tissue cultures for experimentation; the use of polysaccharides to prevent the formation of a glial barrier to nerve growth after injury; observation of changes in reactions of neurons at various stages of development; the finding of hypernatremia and hyperchloremia in lesions of the frontal lobe and the thalamus; stimulation of cerebral blood flow by injection of sodium bicarbonate and retardation with ammonium chloride; and studies of serial sections of brains of palsied children who died.*

*Study of development in the early months of life has made possible the detection of significant abnormalities in behavior early in life. Loss of hearing may be tested in very young children by measuring minute variations in electrical resistance of the skin upon auditory stimulation of the sympathetic nervous system.*

*Conditions which have been described as having been confused with cerebral palsy are dislocation of a cervical vertebra, hereditary spastic paraplegia, transverse myelopathy, injury to the spinal cord or cauda equina by anomalous growths of the spine, and also encephalitis and meningitis.*

*Sedation has proved a valuable adjunct to electroencephalographic study of cerebral palsy. Better criteria for abnormality in the young child should be determined and the application of them more clearly standardized.*

*Simple exercises are useful for early training of palsied children to stimulate development. "Crossed laterality"—the dominant eye being contralateral to the preferred hand—has been counteracted by special training with great success in eliminating emotional and behavior problems and accelerating development.*

*Recent studies indicate that only 50 per cent of cerebral palsy patients have normal or better intelligence.*

*Subluxation of the hip joint, a common deformity associated with cerebral palsy, can sometimes be corrected by operation if detected at an early stage. Radical ablation of epileptogenic foci in the cortex is also being done in young patients if drug control of seizures fails. Frontal topectomy, cingulate gyrectomy or prefrontal lobotomy may be advisable in cases in which proper response to drug therapy is not obtained.*

*Improvement in behavior as well as control of seizures may follow the use of Benzedrine,® Dexedrine,® Dilantin® sodium, Mebaral® and phenobarbital. Alcohol, paraldehyde and chloral hydrate have been effective as relaxants.*

IN REVIEWING BRIEFLY some of the recent work related to cerebral palsy, the author wishes to recommend Sir William Osler's five lectures on "The Cerebral Palsies in Children."[22] Little reference is made today to these excellent articles revealing an understanding of the problem which has not changed too greatly in the succeeding 65 years.

While progress continues in the treatment of cerebral palsy by physical therapy, drugs and surgical procedures, basic research in the pathological factors in this condition offers the best hope for early diagnosis and more effective treatment.

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1. *Basic Research Bearing on Cerebral Palsy*

Pomerat[26] has grown brain cells of various types in tissue cultures as a means of studying the direct effects of many factors in brain cells.

Crude liver or vitamin $B_{12}$ has been found to affect the glial reaction so as to improve the status spongiosus as seen in the lateral columns in Addison's disease.

Permanent damage to the brain due to trauma at birth or afterward may be preventable by medication if the research of Windle, Clemente and Chambers[33] can be successfully pursued. They have been able with the use of a complex polysaccharide, Piromen,® to prevent the formation of a glial bar-

rier to the growth of peripheral nerves in the brain.

Development of brain metabolism studies by Hicks[14] indicates that neurons have widely different reactions at successive stages in development. For example, the neurectoderm is invulnerable to a wide variety of metabolic interruptions. The neuroblast is resistant to anoxia and hypoglycemia but sensitive to sulfhydryl and radiation and the "antisulfyl" action of steroids. Factors that affect glucose metabolism damage the neuron in a newborn child regardless of the oxygen level but, in the adult neuron, the damage is chiefly anaerobic. Interruption of normal steroid metabolism leads to neuron necrosis in the newborn but to marked interstitial and vascular changes in the adult.

In lesions of the frontal lobes and of the hypothalamus, according to Cooper,[8] hypernatremia (with 139 to 155 mg. of sodium per liter in the serum) and hyperchloremia (with 112 to 125 mg. of chlorides per liter in the plasma) usually occur, while the potassium content remains approximately normal. The mechanism causing this imbalance is not understood but may be the cause of some postoperative deaths.

Metabolic alkalosis which causes an increase of 65 per cent in the cerebral blood flow above the resting level may be produced by the intravenous injection of 3 per cent sodium bicarbonate, according to Schieve and Wilson,[29] who also found that a 0.8 per cent solution of ammonium chloride reduced the cerebral blood flow about 25 per cent.

Courville[9] has analyzed residual lesions resulting from antenatal and neonatal asphyxia in an attempt to link them with the abnormalities found in cerebral palsy. Other investigators have questioned whether factors other than anoxia may also be present in birth trauma. Magoun[17] and also Josephy are making studies of serial sections of the brains of children that died with cerebral palsy which it is hoped will lead to better understanding of this condition.

2. *Early Diagnosis*

Gesell and Amatruda[11] have outlined basic patterns of motor, language and personal-social development and by study of infants in the early months of life have been able to detect minor but clinically significant deviations from the norm which became more definite with age. Collis[7] concluded from clinical tests of mental activity in infants that "physical activity of the newborn infant does not display the influence of thought processes, but is entirely reflex . . . the reflex quality normally diminishes as competence emerges, and in the fourth month of life the change can be demonstrated. . . . in full term infants, at the fourth month of life deficient mental maturation is shown by lack of normal subordination of reflex processes; motor dysfunction, on the other hand, is shown . . . by failure of the reflex processes to operate normally in voluntary activity."

Although loss of hearing and loss of vision are frequent in cerebral palsy the early diagnosis of these defects is not easy. Hardy and Pauls[13] have described a "psychogalvanic skin resistance audiometer" developed at the hearing and speech center of Johns Hopkins Hospital. Without previous sedation the subject is seated comfortably and is amused while zinc electrodes are placed on the instep of each foot. Auditory stimulation of the sympathetic nervous system causes sufficient perspiration to reduce the resistance of the skin to the passage of a minute electrical current between the electrodes, from which the changes in voltage are transmitted and recorded.

3. *Differential Diagnosis*

Alexander, Masland and Harris[2] have described a case of anterior dislocation of the first cervical vertebra in which the symptoms of pallor and listlessness occurring at irregular intervals were attributed successively to congestion of the lung, to hypoglycemia, and to birth injury, as also to cerebral palsy.

Spastic paraplegia is another condition difficult to differentiate from cerebral palsy. Ford[10] described hereditary spastic paraplegia as a heredofamilial degenerative disease.

Recently described is transverse myelopathy[23] which resembles thrombosis of the anterior spinal artery but sometimes follows acute infectious diseases, the symptoms including ipsilateral weakness and contralateral loss of pain sensitivity and temperature control without loss of the senses of touch, vibration and position.

Also to be distinguished from cerebral palsy is diastematomyelia caused by the transfixion of the spinal cord or cauda equina by a bony spicule arising from the posterior aspect of a vertebra. In all the cases of this condition reported by Ingraham[16] there was also other anomaly of the spinal column. Neurologic impairment of the lower extremities and of the rectal and vesical sphincters was progressive, and in a number of cases there was a cutaneous midline defect.

The possibility of mistaking encephalitis and meningitis for cerebral palsy is illustrated by the case reported by Benda[4] of a child in whom the only early symptoms were retarded development in sitting, walking and talking, slight strabismus, and "walking with a wide base." Spasticity developed gradually from the age of six years and the child died at 11½ years. "Autopsy showed ample evidence of chronic productive encephalitis which was still in

a progressive state and which involved the cortical gray matter as well as the white matter and basal ganglia. Comparison with lesions in the case of rheumatic encephalitis indicated this diagnosis."

4. *Electroencephalographic Findings*

Of 1,287 children with cerebral palsy in Los Angeles County, 319 were examined by electroencephalogram.[32] Abnormalities were thus indicated in 66 per cent. Of the spastic children, abnormalities were indicated in 70 per cent, 43 per cent of the abnormalities being focal lesions. Abnormalities were likewise indicated in 50 per cent of the children with athetosis, 50 per cent being focal lesions. The percentage of abnormality indicated in "mixed types" was 71, and in children with ataxia, 66.

Ross, Grizzelle and Norfleet[28] studied the electroencephalographic records of 14 children to find whether certain facts of clinical importance could be determined from the records. They concluded that while the electroencephalogram indicates the extent and localization of cerebral lesions this information was not of clinically diagnostic value; intelligence potentials could not be determined from the readings, and prognosis regarding seizures was rarely possible.

In the review by Perlstein, Gibbs and Gibbs[25] of 212 cases of spastic and athetoid cerebral palsy in children, abnormalities were found in the electroencephalograms of 59 per cent of the spastic children who had not had seizures and in 95 per cent of those who had them. In 84 per cent of the spastic children the abnormalities were severe. Abnormalities were indicated in the recordings on 27 per cent of children with athetosis who had not had seizures and in only 67 per cent of those who had. In a series of unselected epileptic children 90 per cent had abnormalities evident in the electroencephalogram, while in normal controls abnormalities were indicated in only 16 per cent. In the recordings of children with cerebral palsy there was a greater frequency of spikes than in the recordings on children with epilepsy, while patterns characteristic of petit-mal and psychomotor disturbances were comparatively rare. They felt that certain inferences could be drawn from such data: "If there has been no history of seizures, the finding of a very abnormal electroencephalogram warns that clinically evident seizures may occur. If there has been no history of seizures the finding of a normal electroencephalogram gives some assurance that clinically evident seizures will not occur."

Aird and Cohen[1] in 1950 reviewed records of 187 cerebral palsy patients between 1 and 21 years of age and studied the electroencephalographic tracings in relation to type and degree of neuromuscular handicap. They concluded that "electroencephalography possesses the same diagnostic and prognostic value in this condition that it does in other neurological disorders associated in a high percentage of cases with cerebral pathologic change and dysfunction. Claims which either belittle its value in this condition or on the other hand stress its unique diagnostic potentialities in cerebral palsy, would appear unwarranted."

Gibbs,[12] at a round table on electroencephalography at the American Academy of Pediatrics meeting in October 1952, described a pattern characteristic of total disorganization (mountainous waves) associated clinically with mass stiffness and spasms. Children in whom these signs were observed almost always died by the age of three years. He stated that if the recording of the brain waves included the period just between sleeping and waking the number of abnormalities found was doubled in cases of epilepsy and increased by a third in cases of psychomotor disturbance. He believed that petit-mal epilepsy could be detected by this method better than by hyperventilation.

Sjaardema[31] has stated that under sedation many abnormalities not otherwise recorded can be detected. He has found that Nembutal,® Seconal,® and paraldehyde often obscure patterns by producing rapid activity. Chloral hydrate gave better results used in 10 per cent solution in syrup, one to four drams orally. He found the best sedative to be Demerol® administered intravenously and followed by sodium pentothal until rapid activity begins. This combination has an effect similar to that of going to sleep and coming out of sleep. Bottle feeding may also induce first alertness and then drowsiness characterized by a pattern of four cycles per second in which focal and unilateral lesions may be detected.

Sjaardema observed that there is no unanimity in the interpretation of electroencephalograms of young children. It would seem that better knowledge of the use of sedation and further review of the criteria for abnormality in the readings are necessary before statistical analyses can have real meaning. For example, of 33 children born with erythroblastosis at Children's Hospital in Los Angeles 25 had some neuromuscular handicap, but in 15 of these children no abnormality was detected by electroencephalogram even though 10 of the 15 were given sedation. Such results do not seem conclusive.

5. *Early Treatment of Neuromuscular Handicap*

A very young infant that does not spontaneously lift his head when in the prone position may be helped to develop this faculty by leaving him on his abdomen for gradually increasing periods of time. Head and trunk control may be developed in a child six to twelve months of age by propping the child in an almost sitting position, asking him to

put out his hands and grasp one's thumbs, and pulling him forward to a sitting position on the edge of his seat, from which he can be assisted to stand on the therapist's lap. A child who does not kick his legs alternately in the normal manner may be trained by exercise of the bicycle type. Many other devices, such as special chairs adapted to particular handicaps and special jumper seats on springs, are being improvised to help the palsied infant to develop at as nearly normal a rate as possible.

6. *Mental and Personality Development*

Berenberg, Byers and Meyers[5] have stressed the importance of promoting maximum personality adjustment as well as physical potential. They point out the well-known socializing influence of the nursery school in its "effect of weaning from close parental control, the relieving of the overtaxed parents and siblings of caring for a child through an unusually long period of dependence," as well as the assistance that can be given the parents by teaching them new methods of dealing with the child's handicap.

McIntire[19] reported that of a series of patients with cerebral palsy 75 per cent were of average intelligence. Hohman,[15] however, reported an exactly reverse proportion in a review at the October 1952 meeting of the American Academy of Cerebral Palsy: 75 per cent of his patients were below average in intelligence, and of these 50 per cent were seriously retarded. Miller and Rosenfield[21] tested 330 children in Buffalo who were under treatment for cerebral palsy and found that 50 per cent had an intelligence quotient below 70, 22.5 per cent were rated between 70 and 89, 23 per cent between 90 and 109, and 4.5 per cent over 110. They found that 40 per cent of the spastic children and the same proportion of those with athetosis were very distractable, and that 40 per cent of the spastic children and 8 per cent of those with athetosis had disturbances of visual control. Of 954 palsied children in Los Angeles County[32] 46 per cent were found to have normal intelligence, only 4 per cent being above average while 21 per cent were on the borderline of normal and 29 per cent had low mentality.

Emotional disturbances, behavior problems in school and retardation in learning have been attributed by Seldowitz and Berman[30] to "crossed laterality"—a condition in which the dominant eye is contralateral to the preferred hand. Special training methods to overcome this handicap have been very successful.

7. *Orthopedic Surgery*

Barnett[3] reviewed orthopedic surgical procedures in relation to cerebral palsy recently and concluded that operation "is only one part of a broad plan of treatment for patients with cerebral palsy; the aim of treatment is to make the patient physically independent and socially competent and acceptable. Although surgery is often required to supplement specific therapies and bracing, it is not to be considered as a substitute for other rehabilitative measures. The value of surgical treatment should not be measured by its aid in overcoming a local deformity or obstruction, but by its contribution to the overall rehabilitation of the patient. A knowledge of proper indications for surgery and adequate preoperative and postoperative care is as essential for good results as a skillful performance of the surgical procedure." Among the common deformities associated with cerebral palsy he included progressive subluxation and dislocation of the hip joint, which was also reported from the Children's Hospital in Los Angeles.[18] If such subluxation seems imminent because of marked adduction deformity, frequent hip x-rays are indicated to detect the earliest stages of subluxation so that surgery can be planned in an effort to prevent further migration of the head of the femur.

8. *Neurosurgery*

Rand,[27] in a recent discussion of the problem and review of the results in the literature, called attention to the fact that in a series of 17 cases of seizures thought to be produced by birth injuries, 76 per cent were relieved from seizures by ablation of the focal epileptogenic lesions of the cortex. Coincidentally with the diminution of seizures, in hemiplegic patients the muscular handicap decreased and behavior improved. Penfield[24] stated that "operative procedures are better controlled and are less subject to risk at 10 years than at 2 but we have been forced to reconsider our policy of postponement in regard to radical ablation of the epileptogenic cortex in those cases in which there is progressive intellectual degeneration or progressive paralysis."

In 40 per cent of palsied children with abnormal behavior electroencephalograms indicate abnormality. Some of these patients respond to drug therapy. Those with most severe emotional disturbance may not, and neurosurgical operation may facilitate general care and rehabilitation. Frontal topectomy, cingulate gyrectomy or prefrontal lobotomy is suggested.

9. *Drug Therapy*

Drugs which reduce or control seizures are important in the treatment of cerebral palsy, since seizures often occur in this condition. For example, in a recent survey in Los Angeles County[32] seizures were noted at some time in 37 per cent of 1,287 cases; 33 per cent of the patients with athetosis, 62 per cent of the spastic children, 40 per cent with mixed symptoms and 42 per cent with rigidity were reported to have had seizures of some kind.

Bradley[6] has pointed out the value of Benzedrine®

and Dexedrine® in behavior disorders in children. Behavior disorders occur very frequently in palsied children, even those in whom no abnormalities are indicated by electroencephalography; these drugs in doses of 5 to 20 mg. a day sometimes afford considerable improvement in behavior.

Improvement in behavior can be obtained in some cases by the use of certain other medication such as Tridione® in doses of 0.3 gm. (5 grains) three times a day or Dilantin® sodium in doses of 32 mg. (½ grain) one to three or more times a day. This improvement may be of a spectacular nature, as in the case of a four-year-old boy who, after study and treatment at the School for Cerebral Palsied Children of Southern California was referred to the local school but could not be managed there. He was given 5 mg. of Benzedrine sulfate daily, and with this medication he has been able to adjust to the local school group.

Alcohol is still probably the best relaxant for cerebral palsy patients, especially those with athetosis, and paraldehyde is sometimes effective. Phenobarbital and Mebaral® have been used with success, the latter especially in tension athetosis in doses up to 0.59 gm. (9 grains) daily. Chloral hydrate in 10 per cent syrup given orally or in 10 per cent aqueous solution given rectally may be used for occasional sedation if other drugs fail to produce sleep.

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